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CASE REPORT

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Spinal cord compression secondary to extramedullary hematopoiesis in thalassemia intermedia

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Abstract Extramedullary hematopoiesis associated with thalassemia causing spinal cord compression is an extremely rare event in the course of the disease. Documentation with an imaging technique, such as MRI, is mandatory. A patient with thalassemia intermedia, who developed paraparesis in spite of transfusion, underwent surgical decompression. Rapid neurological improvement was observed postoperatively and this neurological condition was protected with adequate hemoglobin level. Management of these patients remains controversial. Various modes of therapy such as surgical decompression, radiotherapy, and transfusion are discussed and the related literature is reviewed.

Key words Blood transfusion · Extramedullary hematopoiesis · MRI · Spinal cord · Thalassemia

Introduction

Extramedullary hematopoiesis (EMH) is a common manifestation of severe thalassemia and may involve various organs such as liver, spleen, and lymph nodes [8, 14–16]. It is a compensatory phenomenon that occurs in patients with hematological disorders when the function of the bone marrow is insufficient to meet the demands of the circulation [17]. Spinal cord compression and paraplegia due to extramedullary hematopoietic tissue are extremely rare complications [8, 14, 16, 17]. In 1954, Gatto et al. [4] reported the first case of spinal cord compression secondary to EMH. We present a case of spinal cord compression due to epidural EMH in association with thalassemia intermedia, which was treated successfully with surgical decompression.

Case report

A 37-year-old man with a known diagnosis of thalassemia intermedia, which has previously been treated with splenectomy, presented with a 4-month history of progressive low-back pain with bilateral radiation to the lower extremities. Two months before admission, the patient experienced weakness in his legs. He had a history of back pain and paraparesis, which had improved after transfusion therapy 2 years previously.

On physical examination, the patient was pale, with the characteristic facial features of thalassemia. The liver was palpable at 8 cm below the costal margin. His neurological examination revealed spastic paraparesis with bilateral hypoesthesia below the level of T7. Babinski sign and clonus were positive bilaterally. There was no evidence of sphincter disturbances. Hematological investigation revealed the following: hemoglobin 8 g/dl, hematocrit 26%, white blood cell count 8200/ml, Hb A_2 4.9%, and Hb F 27%. The peripheral smear showed anisopoikylocytosis of red cells with hypochromia, target cell formation, basophilic stippling, 3% nucleated red cells, and 4% reticulocytes. Thoracic MRI revealed a lobulated soft tissue compressing the spinal cord at the level of T4–7 (Fig. 1). The signal intensity of the mass was similar to that of bone marrow and there was no contrast enhancement with gadolinium.

The patient received a total of 1200 ml whole blood over a period of 2 days. After this period, worsening of his neurological condition was noted. He was paraplegic and his sphincter functions were impaired. Urodynamic study showed detrussor sphincter dyssynergia (DSD).

The patient was operated urgently when the paraplegia was noted. After laminectomy, a brownish, soft, lobulated and vascular

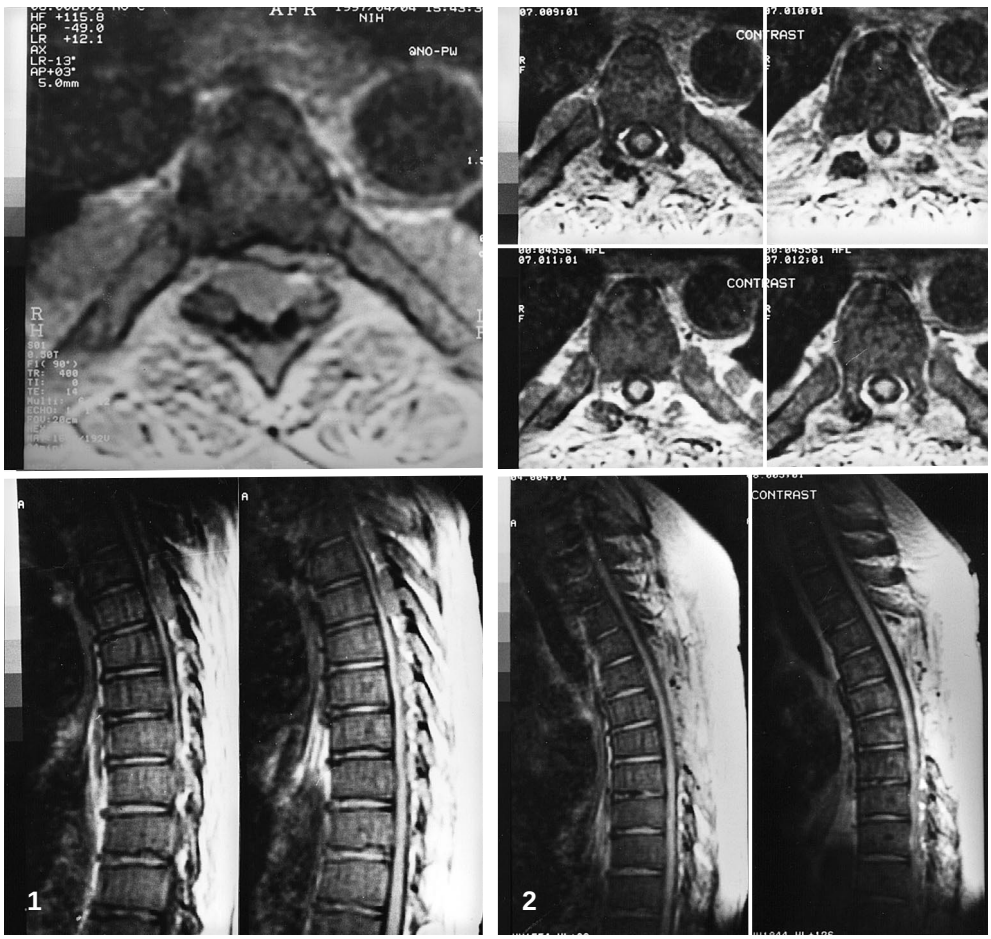

Fig. 1 Sagittal and axial T1-weighted MR images showing a lobulated soft tissue compressing the spinal cord at the level of T4–7

Fig. 2 MR images of the patient 2 months postoperatively showing disappearence of the spinal cord compression

extradural mass extending from T4 to T7 was seen and totally removed.

Histopathological examination of the excised mass and lamina disclosed a hematopoietic tissue. The diagnosis was medullary and extramedullary hematopoiesis.

There was a marked neurological improvement after the operation. Urinary and bowel functions returned to normal and an adequate hemoglobin level was protected by transfusion therapy (hemoglobin 10.2 g/dl, hematocrit 34.4%). The patient was discharged 10 days after the operation. At the follow-up, 6 months postoperatively, his neurological examination revealed no abnormalities. MRI (Fig. 2) and urodynamic studies of the patient disclosed no abnormality.

Discussion

Thalassemia is caused by unbalanced hemoglobin synthesis due to defective production rates of polypeptide chain synthesis. Extramedullary hematopoiesis is a common manifestation in beta thalassemia major and thalassemia intermedia, where it occurs as a compensatory phenomenon in order to combat long-standing anemia [16, 18]. The most common locations of EMH are liver, spleen, kidney, and lymph nodes. Spinal EMH is extremely rare. The development of bone marrow from primitive cell rests or direct extension from the adjacent vertebral bone marrow is the most likely etiology of epidural EMH [1, 3, 7, 8]. There is a predilection for the thoracic spine to cause cord compression [9]; the narrow diameter of spinal canal in this region may be relevant [5, 11, 14, 16].

Spinal cord compression secondary to EMH is a rare neurosurgical disorder. It was first described by Gatto et al. [4] in 1954; since then, a number of cases have been reported in the literature. Singhal et al. [16] compiled 37 cases of spinal EMH and reported a review in 1992.

Clinical awareness is important for early diagnosis. CT can give accurate information about encroachment into the spinal canal of soft tissue masses. Diagnosis can be confirmed via a needle biopsy guided by CT [18]. MRI is the best method for demonstrating spinal cord compression and is an alternative to CT. MRI also appears to be ideal for delineating the extent of the intraspinal masses and visualizes the entire spine [5, 6, 9]; it is thus the modality of choice for the diagnosis and follow-up of these patients [12, 17]. On T1-weighted images, EMH appears as extramedullary masses of signal intensity slightly higher than that of the adjacent bone marrow of the vertebral bodies [9]. Sze at al. advised the restriction of gadolinium enhancement when structural masses cannot

be detected on non-enhanced scans or when meningeal involvement is suspected [19].

There is still considerable controversy regarding the proper management of spinal cord compression in thalassemic patients. Treatment options include surgical decompression, blood transfusions, radiotherapy (RT), or a combination of these modalities [9, 13, 17]. Low-dose RT alone or partial excision and repeated blood transfusion have been reported with good response [8, 9, 13, 14].

It is well known that surgical treatment for spinal EMH is effective. Removal of the epidural mass causes decompression of the spinal cord and rapid neurological improvement occurs. Obtaining a histopathological definitive diagnosis is another advantage of surgical treatment. The disadvantages of surgical therapy include risk of general anesthesia, cardiovascular instability due to anemia, iron overload, and difficulty in total excision due to the diffuse nature of the process [9, 12, 14, 15]. Spinal instability and the risk of reoperation due to rapid recurrence are important postoperative complications. In our case, surgical treatment was found to be beneficial and resulted in immediate neurological improvement. Although the follow-up time is short, the patient has remained symptom free.

Radiotherapy alone is now gradually emerging as the treatment of choice [8, 14, 16, 17]; however, this therapy can not be initiated without a proper diagnosis [15]. Radiation inhibits hemapoietic activity and causes shrinkage of the mass and reduction of the cord compression. According to Singhal et al. [16], the improvement is usually clinically evident after an average of three treatments, and a near complete recovery is generally observed by the end of the treatment. Recurrences at further sites can be managed in similar manner, again with good results. RT is reported to be well tolerated [14]. Cianciulli et al. [2] reported that EMH volume decreased by 16.4% immediately after RT.

Some authors advise that transfusion therapy should be regarded as the initial form of management in thalassemic patients with spinal cord compression due to EMH [3]. With blood transfusions, the need for EMH decreases, and this will result in the relative inactivating of those tissues, which revert to their initial size. However, improvement is usually incomplete and the symptoms recur shortly after. Blood transfusion will have no effect on edema or cord compression. It can, however, be used both as a diagnostic as well as a therapeutic method to obviate the need for surgery or RT in mild spinal cord compression [10, 13, 15, 16]. Transfusions have been used both as a sole treatment and as an adjunct to surgery and to RT [13]. In our patient, paraparesis had improved after transfusion therapy at first admission in 1995, but 2 years later recurrence occurred and no improvement was observed with blood transfusion. The patient was therefore operated and the paraparesis disappeared.

In our opinion, surgery is the treatment of choice in the event of acute and severe neurological deficits. RT or transfusion therapy may be regarded as the initial management in cases with mild paraparesis, especially in patients who have a known history of thalassemia. The use of these treatment modalities, on their own or in combination, depends on the clinical condition of the patient.

Conclusion

Spinal cord compression secondary to EMH is a rare neurosurgical entity. Treatment options include surgical decompression, RT, transfusion, and any of these in combination. Proper management depends on the neurological status of the patient. While surgical decompression is necessary in patients who have progressive neurological deficit or acute paraplegia, transfusion therapy may be useful as a diagnostic as well as a therapeutic method in moderate spinal cord compression secondary to EMH. Surgery and RT can be reserved for those patients if transfusion alone fails.

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